COR BILOCULARE

BY

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A two-chambered heart is a rare congenital anomaly. The following case survived to the fifteenth week of life without any obvious signs.

Case History

The infant was the fourth child of healthy English parents and there was no known history of congenital anomalies in any other members of the family. The mother's pregnancy was normal apart from mild hypertension (B.P.=140/90). Her blood group was B Rhesus negative (cde/cde) and no antibodies were detected in her serum by the antiglobulin test at 36 weeks.

A spontaneous vertex delivery of a female infant, whose birth weight was 4 lb. 14 oz. occurred at 37 weeks. Clinical examination was normal and no cardiac murmurs were detected. The infant was breast-fed until the eighth week of life when a change to bottle feeding was made. Her progress was satisfactory and at 14 weeks she weighed 9 lb. 5 oz. The parents did not notice any dyspnœa and only in retrospect did they comment that the infant may have become slightly blue when she cried.

In the fifteenth week of life the child developed a mild upper respiratory infection. Three days later she suddenly became cyanosed shortly after a feed, and her breathing became irregular. She quickly lost consciousness and death occurred within ninety minutes of the onset of the attack.

Necropsy. The body was that of a small but well-nourished female infant with arachnodactyly and a high arched palate. Complete transposition of the viscera with dextrocardia was present. The heart was enlarged, weighing 65 g. with a transverse diameter of 5 cm. (Fig. 1). A single superior vena cava and two main pulmonary veins opened into the posterior aspect of a single atrium. The inferior vena cava could not be traced to the heart, and the azygos vein carried the venous blood from the lower part of the body. A tiny auricular remnant measuring $\frac{1}{2} \times 1$ cm. was present in the atrio-ventricular groove on the right side, but had no demonstrable communication with the interior of the heart. The atrium communicated with a common ventricle through a single tricuspid A-V valve 2 cm. in diameter. The posterior cusp was considerably smaller than the others. The ventricular muscle was moderately hypertrophied, its thickness varying from 7 to 14 mm. (Fig. 2). The aorta, the aortic valve, and the origin and distribution of the coronary vessels were normal. The pulmonary trunk was a mere fibrous remnant but the main branches were dilated and communicated with a widely-patent ductus arteriosus.

Histologically, the heart muscle appeared normal. The lungs showed widespread areas of collapse with a few areas of emphysema: large numbers of macrophages were present in the alveoli and there was lymphoid hyperplasia. The aorta showed no evidence of medial degeneration.

Discussion

Cor biloculare is probably the rarest of all congenital malformations of the heart (Wood, 1956). Abbott (1936) found only 14 cases of a two-chambered heart in 1000 cases of congenital heart disease.

Brown (1950) divided the condition into three groups: (1) with an undivided truncus arteriosus, (2) with normal division into aorta and pulmonary artery, and (3) incomplete forms with some septal formation but a persistent single A-V valve. Our case appears to belong to the second group. Cor biloculare is usually classified with the cyanotic group of congenital heart diseases (Bedford and Brown, 1951). Sometimes, however, as in this instance, there must be minimal mixing of venous and arterial blood streams within the heart, with little or no cyanosis (White, 1951), and the absence of murmurs and thrills makes the diagnosis

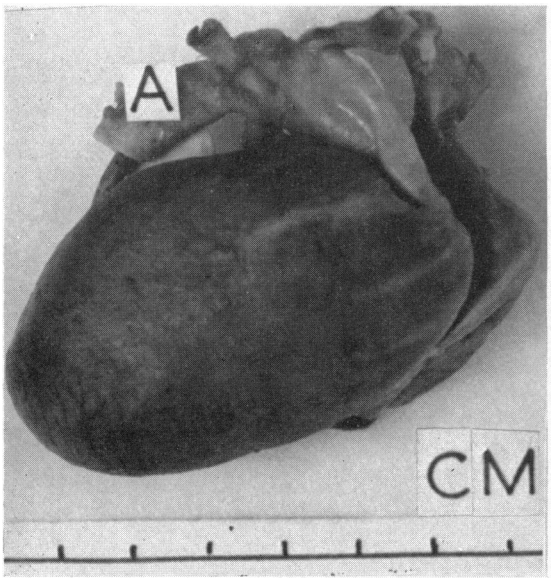

FIG. 1.—The heart as it was *in situ*, showing cardiac enlargement and dextroposition of the aorta (A).

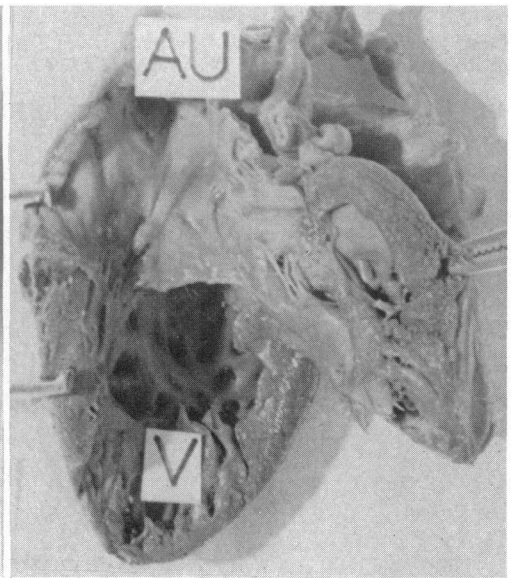

FIG. 2.—The opened heart viewed from the back looking towards the left. It shows the single atrium (AU) and ventricle (V), which is hypertrophied. The scale is the same as in Fig. 1.

in life extremely difficult. Campbell *et al.* (1952) reported a case of cor biloculare with isolated lævocardia and an absence of the inferior vena cava also: even after full investigations the exact diagnosis was made only post mortem, although the authors stated in retrospect that some of the X-ray and angiocardiographic evidence might have suggested the diagnosis. At present it seems that a confident diagnosis of cor biloculare will seldom be made in life: White (1951) does not include the condition in his classification of diagnosable congenital defects of the heart.

The survival of patients with such a gross cardiac defect is surprising. Abbott (1936) gave the mean age at death as 3¼ years, the oldest of her series surviving to 16 years. Nelson and Wells (1948) have described a patient with cor biloculare who lived 27 years.

In two-chambered hearts a single A-V orifice is almost always present, but Taussig (1947) has described a case in which two A-V orifices were present. The A-V valve usually has three or four cusps but it is sometimes difficult to be sure of the exact number and the arrangement of the chordae is often unhelpful. Schechter and Moranze (1944) have reported a case of cor biloculare associated with dextrocardia in which the A-V valve consisted of one small cusp and two large ones, an arrangement very similar to that in our case.

Cor biloculare hardly ever occurs without other anomalies being present (Brown, 1950) and these were abundant in the case described here. Dextrocardia, varying degrees of situs inversus and other visceral anomalies are common associated findings. The presence of arachnodactyly and a high arched palate in this case was suggestive of Marfan's syndrome, but there were no other skeletal, ocular, or aortic abnormalities to support this diagnosis.

Summary

An infant with cor biloculare associated with arachnodactyly and complete situs inversus, who survived to the fifteenth week of life, is described. Attention is drawn to the lack of physical signs despite the profound abnormality. The extreme difficulties in diagnosis are discussed.

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CORRIGENDUM

In the advertisement by Laboratory Nativelle Limited, published in the November, 1961 issue of the BRITISH HEART JOURNAL, it was stated in error that tablets of 0.1 m.g. were "1/6000 gr." The advertisement should have read "Tablets 0.1 mg. (1/600 gr.)."